A 38-year-old man presented with a 3-month history of night sweats, nonproductive cough, and weight loss. He denied chest pain or shortness of breath. A transesophageal echocardiogram showed a large mass adjacent to the aortic root likely arising from the pericardium or the left ventricle (Figure 1 and Movie I in the online-only Data Supplement). Invasive coronary angiography revealed a highly vascular mass perfused by the left main artery causing deformity of the aortic root (Figure 2 and Movie II in the online-only Data Supplement).

Various imaging modalities were then performed to further characterize the lesion. Cardiac computed tomography angiography revealed a large heterogeneous, intensely enhancing mass within the pericardial sac, just above the anterior wall of the left ventricle, anterior to the left atrium and to the left of the ascending aorta. The mass was encasing the left main artery and the proximal left anterior descending artery (Figure 3).

Cardiac MRI revealed a large, well-circumscribed mass arising from the pericardium in the left paraaortic region. The mass was isointense to myocardium on T1-weighted images (Figure 4A) and highly hyperintense on T2-weighted images (Figure 4B). It did not demonstrate features to suggest invasion of the adjacent myocardium or vascular structures. Rather, it appeared to be displacing structures favoring a benign entity (Movie III in the online-only Data Supplement).

Computed tomography scans of the abdomen, pelvis, and head confirmed the absence of distant metastases.

Because the patient was hypertensive, a 24-hour urinary collection was performed, which demonstrated high levels of normetanephrine and metanephrine that are suggestive of a pheochromocytoma. Two days later, a retrograde transvenous venous catheter collection was performed, which demonstrated high levels of normetanephrine and metanephrine that are suggestive of a functioning paraganglioma (Figure 4B). It demonstrated that the mass was isointense to myocardium on T1-weighted images and hyperintense on T2-weighted images (Figure 4C). It did not demonstrate features to suggest invasion of the adjacent myocardium or vascular structures. Rather, it appeared to be displacing structures favoring a benign entity (Movie III in the online-only Data Supplement).

Paragangliomas are rare tumors of chromaffin cells arising from an extra-adrenal location. The majority of these tumors originate from the adrenal medulla and are known as pheochromocytomas. Intrapерicardial paragangliomas are extremely rare and have been reported adjacent to the bifurcation of great vessels. They tend to be located in the aortopulmonary groove, close to the left atrium, at the level of the para-aortic sympathetic ganglia. The presenting symptoms are variable and may be related to catecholamine hypersecretion with hypertension, palpitations, headache, and sweating or may be attributable to the mass effect on adjacent structures. In some cases, when the tumor is nonfunctioning, they may present as an incidental finding.

Intrapерicardial paraganglioma should be considered in the differential diagnosis of a well-circumscribed, highly vascular mass arising from the aortopulmonary groove demonstrating high signal intensity on T2-weighted sequences on MRI and intense contrast enhancement on computed tomography. The differential diagnosis should include other pericardial tumors, namely pericardial hemangiomata, sarcoma, and metastases. Although imaging findings together with the presence of hypertension in a patient with catecholamine excess strongly suggest the diagnosis, tissue diagnosis is ultimately needed. Multimodality imaging plays an important role in planning a complete surgical resection, which is the curative treatment.

Disclosures

None.

References


Figure 1. Transesophageal color Doppler echocardiogram demonstrates that the left coronary artery (arrow) was embedded in a large solid mass (M) that was perfused by small branches from the same artery.

Figure 2. Selective coronary angiogram of the left coronary artery demonstrates multiple feeding vessels extending into a mass.

Figure 3. Cardiac computed tomography (CT) of intrapericardial paraganglioma. Coronal oblique multiplanar reconstruction of a contrast-enhanced cardiac CT angiogram, demonstrates a heterogeneous, avidly enhancing mass (M) located between the ascending aorta (Ao), the pulmonary artery (PA), and the base of the left ventricle. Note the left main coronary artery traversing the mass (arrow).
Figure 4. A, Cardiac magnetic resonance images of intrapericardial paraganglioma. Axial T1-weighted Fast Spin Echo image demonstrates an isointense mass with multiple vascular channels, seen as signal voids due to rapidly flowing blood (arrow). Note the pericardium (arrowhead) with the overlying epicardial fat surrounding the anterolateral aspect of the mass, confirming its intrapericardial location. B, Cardiac magnetic resonance images of paraganglioma. Axial T2-weighted Fast Spin Echo Fat Saturation image demonstrates a hyperintense mass containing multiple vascular channels (arrow). The mass is adjacent to the ascending aorta and splaying the right ventricular outflow tract (RVOT) and left atrium (LA).

Figure 5. Paraganglioma. A, Tumor displaying mixed nested (zellballen) and trabecular patterns (hematoxylin-eosin stain, magnification ×100). B and C, Immunohistochemistry for neuroendocrine markers shows that the tumor cells are strongly positive for chromogranin A (B) and synaptophysin (C; magnification ×100). D, Gross image of the surgical specimen showing a well-circumscribed, lobulated lesion with a brown-yellow-red cut surface. Note the dilated vascular channels within the mass (arrows). E, The sustentacular cells are highlighted by S100 (immunohistochemical stain, magnification ×200).
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MacArinze Ojiaku, Elena Peña, Eric C. Belanger, Kwan-Leung Chan and Carole Dennie

Circulation. 2014;130:e137-e139
doi: 10.1161/CIRCULATIONAHA.114.012458

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